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CASE REPORT

Co-occurrence pulmonary haemosiderosis with coeliac disease in child

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Summary
Idiopathic pulmonary haemosiderosis (IPH) is a rare and serious disorder in children of unknown aetiolopathogeny. Association of IPH and coeliac disease (CD) is even rarer. Immunological origin of IPH is now well accepted. We report the case of an 11-year-old female admitted for evaluation of recurrent streaky haemoptysis that had been evolving over the previous 9 months. Physical examination revealed weight loss with normal weight, but there was cutaneous and mucosal pallor due to severe anaemia (haemoglobin 4.6 g/dl). The chest X-rays showed unilateral alveolo-intertitial infiltrate. Broncho-alveolar lavage revealed 70% haemosiderin-laden macrophages. The diagnosis of IPH was made. Since severe anaemia is disproportionate to radiologic findings, searching associated CD was performed and then confirmed by biological and histological examinations. A gluten-free diet was initiated. Evolution was favourable. Looking for especially CD in IPH should be systematic, even in the absence of gastrointestinal symptoms.

Introduction

Idiopathic pulmonary haemosiderosis (IPH) is a rare disorder in children, characterized by a triad of recurrent episodes of alveolar haemorrhage, haemoptysis and anaemia. If untreated, prognosis is poor because of fibrosis and restrictive lung disease.[1] The association of IPH and coeliac disease (CD) has been rarely described, underlining a close aetiopathogenic link between both diseases. We report a new paediatric case of IPH associated with CD.

Case report

An 11-year-old female presented to our paediatric unit on the 13th of March 2006 for tachypnea and fever. The infant had a 9-month's history of streaky haemoptysis. She weighed

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33 kg and her height was 1.34 m, temperature was at 38.2 °C. Physical examination noted a marked pallor, polypnoea, tachycardia and a systolic heart murmur without signs of heart failure. Blood pressure was 9/5. The remaining physical examination was normal. The infant had no signs of malabsorption.

Complete blood count showed severe microcytic hypochromic aregenerative anaemia (Hb 4.6 g/dl) for which she received two units of packed red cells. Iron investigation showed hyposideraemia at 2 μmol/l, total capacity of serum fixation at 73.2 μmol/l and transferrin saturation coefficient at 2.7%, compatible with iron deficiency. Haemoglobin rate achieved 12 g/dl after transfusion. Laboratory tests, including renal function, serum electrolytes, urine examination and liver enzymes were normal. Inflammatory markers were slightly increased.

Pulmonary tuberculosis or pulmonary haemosiderosis were evoked. Tuberculin's intradermo-reaction and research of Kenoch Bacillus in gastric tubage were negative, excluding diagnosis of tuberculosis.

As diagnosis of IPH was considered, bronchoalveolar lavage was preformed. Bronchoalveolar lavage confirmed diagnosis, based on the presence of 70% haemosiderin-laden macrophages.

A chest X-ray revealed unilateral alveolo-intertitial infiltrate. Spirometry was normal. Echocardiography demonstrated left ventricle dilatation, secondary to severe anaemia, with normal pulmonary pressure. Coagulation studies, including Von Willebrand's factor, were normal. Serum antinuclear antibody and antiglomerular basement membrane antibody were negative. Serum Immunoglobulin and complement rate were normal. Since the association between CD and IPH has been previously reported, and looking for severe anaemia disproportionate to radiologic findings, investigations for CD were performed. Serum IgA antiendomysial and ant tissue transglutaminase antibodies were positive at a titre of more than 100 UI/ml (normal: 0–10 UI/ml). A duodenal biopsy revealed total villous atrophy consistent with a diagnosis of CD. The Infant received a gliadin-free diet and iron supplementation. Clinical course improved without recurrence of haemoptysis over 12-month follow-up. Haemoglobin at 12-month follow-up is 12.8 g/dl, antiendomysial antibody titres have declined to 20 UI/ml, indicating compliance with gliadin-free diet.

Discussion

IPH is a rare disease of unknown aetiology, usually seen in childhood. Incidence varied from 0.24 to 1.23 patients per million children.[2,3] The triad of anaemia, haematemesis and recurrent alveolar haemorrhage suspects diagnosis and the finding of haemosiderin-laden macrophages in bronchoalveolar lavage fluid confirms diagnosis.[4] In infants with IPH, the co-occurrence with CD has been described, underlining the importance of autoimmune phenomena in this disorder. The combination of IPH and CD has been rarely reported in literature.[5] At diagnosis, age varied from 2 to 56 years and gliadin-free diet improves pulmonary disease. Currently, children with CD manifest clinical signs of malabsorption with significant diarrhoea, steatorrhea, impaired growth, abdominal distension and muscle wasting.[6] Our patient had no signs of classic or non-classic CD. She had only severe iron-deficient anaemia. For this and since CD had been previously reported in association with IPH, serological test and intestinal biopsy were performed and confirmed diagnosis of CD. The patient was started on a gluten-free diet with iron supplementation. She has had no recurrence of haemoptysis over a follow-up of 12 months. Actually, it is admitted that CD and IPH are both immunologically mediated diseases and may have a common pathogenetic link.[7] The pathogenetic link between these diseases is still unclarified.[8] In IPH, several mechanisms can damage pulmonary capillaries such as antibodies to type IV collagen occurring in Good pasture syndrome, deposition of immune complexes in autoimmune disease with activation of complements cascade leading to membrane damage.[5,6] In CD, an important fact is that CD results from an inappropriate T-cell-mediated immune response against gluten in genetically predisposed people.[9] The link between CD and IPH is important to recognize because previous reports suggest that treatment of CD may lead to remission of IPH.[8,10] Our patient has remained well without recurrence of haemoptysis on a gluten-free diet for 12 months.

Conclusion

To conclude, our case shows that CD should be specifically looked for in patients with IPH, especially those in whom the severity of anaemia is disproportionate to radiologic findings despite the absence of digestive symptoms since both diseases may benefit from gluten-free diet.

Conflict of interest statement

None of the authors have a conflict of interest to declare in relation to this work.

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